Musculoskeletal Complications of Hemophilia

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• **Persons with hemophilia may be crippled by hemorrhages into soft tissue, the pressure destroying nerve and muscle. Recurrent bleeding into joints produces severe arthritis with synovitis and damage to cartilage and bone. The resulting deformities, even of long standing, may be greatly lessened, so far as impairment of function is concerned, by conservative orthopedic treatment. Treatment also may slow the progress of crippling deformities.**

MANY PHYSICIANS rarely see hemophiliac patients. When they do, they apparently tend to undertreat them, lest they do more harm than good. This paper presents nothing new, but is a review of musculoskeletal problems associated with hemophilia that have been observed by the author and a report on how these problems were dealt with. The joint destruction, the contractures and the paralysis of hemophiliac patients resemble arthritis, club feet and poliomyelitis and are often amenable to the same care. Surgical treatment, of course, is out of the question, but conservative orthopedic measures are sometimes a boon.

Several investigators—Ghormley and Clegg,[6] Thomas,[14] Caffey and Schlessinger[2] and others*—have written excellent reviews of the problem of hemophilic arthritis, but that their work may not be sufficiently well known is indicated by the number of patients observed with crippling deformities which have never been treated, although they might have been held to lesser degree had they been dealt with in the early stages. Indeed, many such patients have improved significantly even with delayed care.

There are 242 hemophiliac persons registered with the Southern California chapter of the Haemophiliac Society. One hundred and ten, or 45 per cent, are patients with musculoskeletal disorders resulting from the disease. It is with them that this paper is concerned. With the exception of one case of paraplegia, which occurred subsequent to a spinal puncture, I have not seen involvement of the spine, and my experience has been entirely with disorders of the extremities.

The problems are both acute and chronic, the latter being the more difficult to manage over a long period.

ACUTE PROBLEMS

Hemorrhage

Control of bleeding by reducing the clotting time is an emergency measure.[16] Hemorrhage can arise spontaneously as well as from various degrees of trauma. Hematologists do not agree upon details of measures necessary to reduce the clotting time. In the majority of cases I administer Hyland Laboratories' antihemophilic plasma in a dose of 1 to 2 cc. per pound of body weight. In 1956, 2,041 liters of plasma and 174 units of whole blood were administered to hemophiliac persons registered with the Haemophiliac Society in our area, including those with visceral bleeding. Plasma is supplied without charge to all patients. Those living at a distance keep it in their refrigerators for administration by a local physician.

Fractures

Fractures are treated by the orthodox methods of traction, splinting and casting. Sometimes breaking a bone causes more bleeding in a hemophiliac than in a normal person, but usually the hematomas are not large as compared with some of the larger ones that occur in soft tissue after considerably less trauma than that needed to break a bone. Theoretically, traumatic damage to tissue engenders a substance necessary in clotting.[5]

Hematomas associated with fractures in hemophiliac patents should not be aspirated or evacuated, for the puncture or incision would be hazardous. Circular casts are not applied until swelling has subsided and early healing of the bone is established. Open reduction is out of the question; deformity is more acceptable. Bone healing occurs at a normal rate, usually with abundant callus.

Soft Tissue Bleeding

Bruises often remain small but can grow insidiously into enormous crippling hematomas, and each one must be watched. Patients gain experience with their own bleeding problems, which differ in each case and at various times. They often can forecast quite accurately the likely extent of a hemorrhage.

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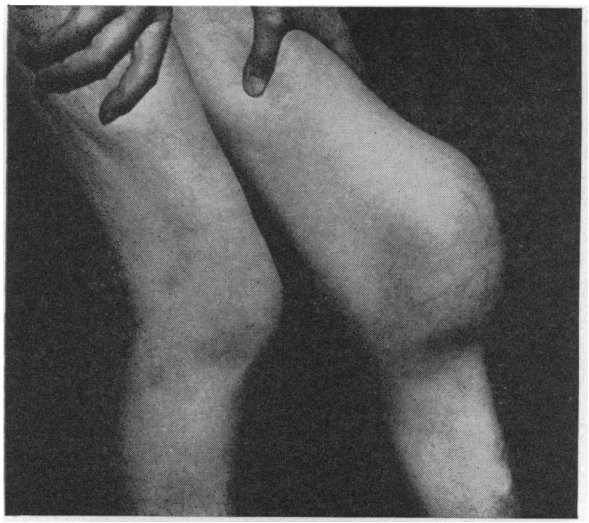

Figure 1.—Swollen knee resulting from recurrent intra-articular hemorrhages. Note striae.

It is to be emphasized, however, that, when doubt exists, efforts to stop the bleeding should be started immediately. In addition to lowering the clotting time by the administration of antihemophilic plasma, these measures consist of absolute rest of the part and packing the area of hemorrhage in ice. This is best done in the hospital where the swelling can be observed and nursing care and drugs are available for control of pain and restlessness.

Bleeding into Joints

Minor leakage of blood into a joint is rare. When hemorrhage develops in a joint the joint usually distends to full capacity until the intra-articular pressure is as high as the systolic pressure and no more blood can be pumped in. More may leak in, however, as the initial hemorrhage absorbs, a process which may continue indefinitely.[8] Patients with intra-articular hemorrhages should be put in hospital for care in order to minimize permanent damage (see Figure 1). Bleeding into a joint is usually very painful, a symptom useful to distinguish it from the lesser discomfort of the relatively less serious bleeding adjacent to a joint.

Deformity also requires treatment. I use plaster shells for elbows, wrists and ankles, splinting the elbow at a right angle, holding the wrist in the position of function and splinting the ankle at a right angle to prevent foot drop. For hip and knee joints, Russell traction or Buck's extension is used, the patient wearing a padded tennis shoe. Traction should not be used to try to correct the contracture but should pull in the line of the deformity and be adjusted daily as muscle spasm subsides.

Even though recognizing that the presence of free blood in a joint may do severe damage to it, I consider aspiration in such circumstances unwise, for the trauma of the needle may produce further bleeding even after the clotting time has been brought to normal. I have not had experience in the injection of hyaluronidase in an effort to speed absorption of blood.[1,9]

Addiction to Narcotics

As with many chronic diseases, the danger of addiction is always a factor in dealing with the painful concomitants of hemophilia. Many hemorrhages, particularly intra-articular, are so painful as to make the use of narcotics necessary for relief. Some of the patients I have treated were already addicted when first observed, narcotics apparently having been the only previous treatment.

THE PROBLEM OF CHRONIC DISABILITY

The chronic disabilities of hemophilia—muscle weakness and paralysis, contractures, synovitis and degenerative arthritis — are a major orthopedic problem.

Weakness and Paralysis

Muscle weakness arises either because of nerve damage resulting from pressure of a hematoma in a closed fascial space or from disuse of an extremity. I treat it as though it were the muscle weakness of poliomyelitis, using electrical stimulation and exercises to restore strength, and braces for support and the control of antagonists. The quadriceps is the most commonly weakened muscle, usually as the result of chronic hemophilic arthritis in the knee joint but sometimes because of damage to the femoral nerve by groin hemorrhage. Volkman's contracture and peroneal palsy sometimes occur.

Contractures

Contractures of hips, knees, feet and ankles respond well to traction, braces and, occasionally, wedging plasters, along with muscle reeducation. Gentleness is essential. These joints will bleed if quickly forced. The knee is the most frequently involved (Figure 2). I have used adjustable long-leg braces successfully to overcome knee joint flexion contractures of long standing. The patient is permitted to walk despite the deformity, but a posterior bar on the brace blocks further flexion while the leverage of the brace gently urges further extension with each step. As the contracture diminishes, the brace is adjusted gradually until the knee reaches full extension, when free motion is started.

Improvement can be brought about in "frozen" shoulders by using the customary measures of x-ray therapy to control pain, then heat, massage, stretching and exercise.

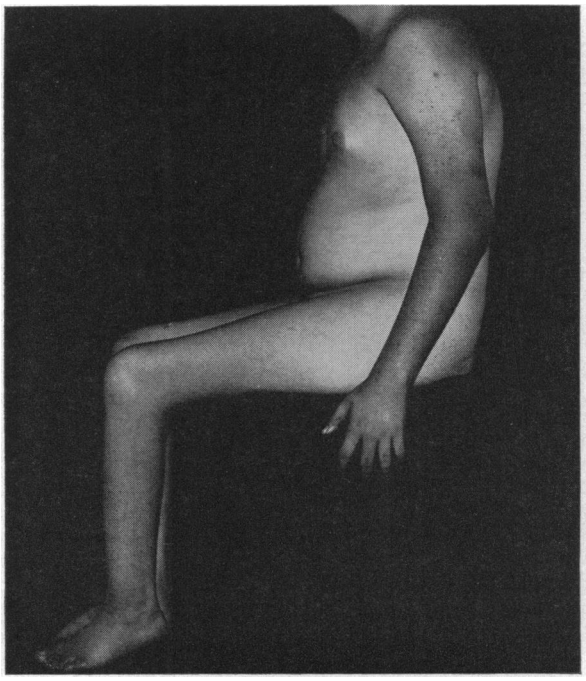

Figure 2.—Fixed knee flexion contractures resulting from untreated recurrent joint bleeding.

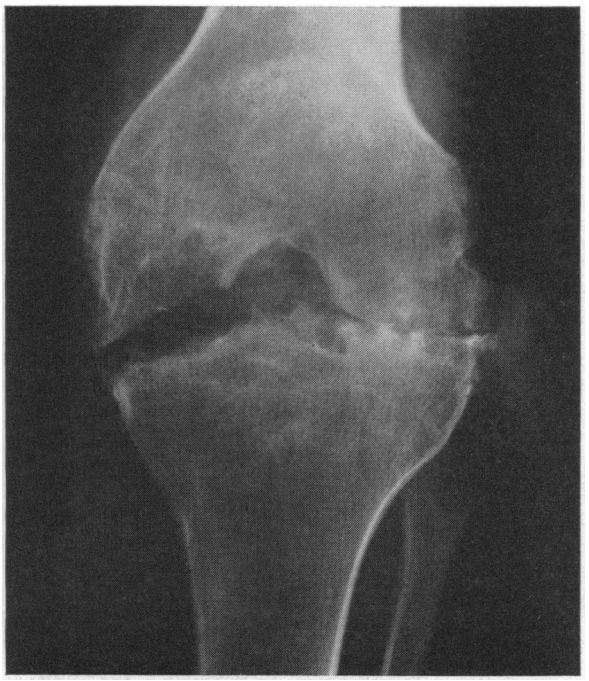

Figure 3.—X-ray film showing advanced hemophilic arthritis of knee in a 14-year-old boy, characterized by erosion articular surface, lipping of joint margins, subchondral bone cysts.

Synovitis

Unfortunately, the majority of contractures are complicated by various degrees of synovitis and articular damage resulting from a series of hemorrhages. The prognosis for full recovery of these joints is poor, but worthwhile functional improvement can be obtained.

Experimental evidence has been reported that synovial hyperplasia[10] and subsequent hemovillous villonodular synovitis[18] can result from repeated hemorrhage. Hyperplastic synovitis can be reduced, but treatment has been disappointing in the few patients suspected of having villonodular synovitis with whom I have dealt. Treatment of arthritic joints is the treatment of the contractures plus an attack on the synovitis. For this I use hydrocortone intra-articularly, x-ray therapy and prednisone (Meticorten®) by mouth. The number of patients so treated is too small for statistical conclusions, but the results of all three forms of treatment have been encouraging.

Hydrocortone is injected intra-articularly, after the clotting time has been reduced to normal by infusion of plasma. This produces significant relief of pain and clinical improvement in synovial thickening and fluid. It has not restored a joint to normal in any case observed.

X-ray therapy has produced varying degrees of improvement in about 80 per cent of patients with chronic synovitis. It has been particularly helpful in reducing the swelling and pain and increasing the range of motion in elbow joints. The clinical improvement, however, has not always been as evident as the patient's gratifying relief from pain. Lest epiphyses be damaged, x-ray therapy has been reserved for adults only.

I have used oral Meticorten in 14 patients with multiple joint involvement. All of them have reported decided diminution of pain and improvement in their sense of well-being. Moderate improvement in the joints has been observed clinically. How long Meticorten therapy can be continued and precisely what the dose should be cannot be said as yet.

Osteoarthritis

Although we have no method of direct attack on damage already done to bone and cartilage,[3] treatment of weakness, contractures and synovitis often can restore a badly damaged useless joint to useful function. I believe treatment of this kind also can decrease the rate of further degeneration. I believe that treatment of the acute episodes of hemorrhage and care of the deformities will at least postpone crippling.

The roentgenographic appearance of osteoarthritis (Figure 3) sometimes resemble the changes seen in rheumatoid arthritis and tuberculosis. Typically, there is narrowing and roughening of the joint spaces, increased opacity of the soft tissues due to deposition of iron pigments, osteoporosis, epiphyseal cysts and spurring at the epiphyses.

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